

Serial No. 10/826,112
Atty. Docket No.: P71641US0

Listing of Claims:

1. (Currently amended) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement in which the weight ratio of Leu to Val is greater than 2:1; and

enterally administering to the subject a said LNAA supplement ~~in which the weight ratio of Leu to Val is greater than 2:1.~~

2. (Currently amended) A The method according to claim 1, wherein the LNAA supplement is substantially free from phenylalanine.

3. (Currently amended) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement in which the weight ratio of Leu to iLeu is greater than 3:1; and

enterally administering to the subject a said LNAA supplement ~~in which the weight ratio of Leu to iLeu is greater than 3:1.~~

4. (Currently amended) A The method according to claim 3, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

5. (Currently amended) A The method according to claim 4, wherein

the LNAA supplement is substantially free from phenylalanine.

6. (Currently amended) A method for treating a subject suffering from phenylketonuria and/or phenylalanemia, said method comprising the steps of:

obtaining a LNAA supplement which comprises one or more LNAAs and which further comprises Lys; and

enterally administering to the subject a said LNAA supplement~~which comprises one or more LNAAs and which further comprises Lys.~~

7. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu.

8. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1.

9. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

10. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

11. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu, and wherein the weight ratio of

Leu to Val in the LNAA supplement is greater than 2:1.

12. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu; wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1; and wherein the weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

13. (Currently amended) A The method according to claim 12, wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

14. (Currently amended) A The method according to claim 12, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

15. (Currently amended) A The method according to claim 6, wherein the LNAA supplement comprises Leu; ~~wherein the~~ said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1; and ~~wherein the~~ said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

16. (Currently amended) A The method according to claim 6, wherein the LNAA supplement is substantially free from phenylalanine.

17. (Currently amended) A LNAA supplement comprising Leu and Val, ~~in which~~ wherein the weight ratio of Leu to Val is greater than 2:1.

18. (Currently amended) A The LNAA supplement according to claim

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17, wherein the LNAA supplement is substantially free from phenylalanine.

19. (Currently amended) A LNAA supplement comprising Leu and iLeu, in which the weight ratio of Leu to iLeu is greater than 3:1.

20. (Currently amended) A The LNAA supplement according to claim 19, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

21. (Currently amended) A The LNAA supplement according to claim 20, wherein the LNAA supplement is substantially free from phenylalanine.

22. (Original) A LNAA supplement comprising one or more LNAAs and further comprising Lys.

23. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement further comprises Leu.

24. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu and ~~wherein the~~ said weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1.

25. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu and ~~wherein the~~ said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

26. (Currently amended) A The LNAA supplement according to claim

22, wherein the LNAA supplement comprises Leu, and ~~wherein the~~ said weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

27. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu, and ~~wherein the~~ said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

28. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu; ~~wherein the~~ said weight ratio of Leu to iLeu in the LNAA supplement is greater than about 0.5:1; and ~~wherein the~~ said weight ratio of Leu to Val in the LNAA supplement is greater than about 0.5:1.

29. (Currently amended) A The LNAA supplement according to claim 28, wherein the weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1.

30. (Currently amended) A The LNAA supplement according to claim 28, wherein the weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

31. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises Leu; ~~wherein the~~ said weight ratio of Leu to iLeu in the LNAA supplement is greater than 3:1; and ~~wherein the~~ said weight ratio of Leu to Val in the LNAA supplement is greater than 2:1.

32. (Currently amended) A The LNAA supplement according to claim

22, wherein the LNAA supplement is substantially free from phenylalanine.

33. (Currently amended) A The LNAA supplement according to claim 22, wherein the LNAA supplement comprises, per 500 mg of LNAA supplement:

| | | | | |
|------------|-----|-------------|-----|----------------|
| from about | 100 | mg to about | 290 | mg of Tyr; |
| from about | 25 | mg to about | 75 | mg of Trp; |
| from about | 15 | mg to about | 50 | mg of Met; |
| from about | 15 | mg to about | 55 | mg of iLeu; |
| from about | 15 | mg to about | 50 | mg of Threo; |
| from about | 15 | mg to about | 55 | mg of Val; |
| from about | 15 | mg to about | 200 | mg of Leu; |
| from about | 10 | mg to about | 30 | mg of His; and |
| from about | 5 | mg to about | 200 | mg of Lys. |

34. (Currently amended) A The LNAA supplement according to claim 33, wherein the LNAA supplement comprises, per 500 mg of LNAA supplement, ~~from~~ about 10 mg to about 30 mg of Lys.

35. (Currently amended) A The LNAA supplement according to claim 33, wherein the LNAA supplement is substantially free from arginine.

36. (Currently amended) A The LNAA supplement according to claim 33, wherein the LNAA supplement is substantially free from phenylalanine.

37. (Original) A LNAA supplement comprising, per 600 mg of LNAA

supplement:

from about 100 mg to about 290 mg of Tyr;
from about 30 mg to about 90 mg of Trp;
from about 25 mg to about 75 mg of Met;
from about 15 mg to about 45 mg of iLeu;
from about 15 mg to about 50 mg of Threo;
from about 15 mg to about 50 mg of Val;
from about 40 mg to about 200 mg of Leu;
from about 15 mg to about 45 mg of His; and
from about 15 mg to about 50 mg of Arg.

38. (Currently amended) A The LNAA supplement according to claim 37, wherein the LNAA supplement further comprises Lys.

39. (Currently amended) A The LNAA supplement according to claim 37, wherein the LNAA supplement further comprises, per 600 mg of LNAA supplement, ~~from~~ about 5 mg to about 200 mg of Lys.

40. (Currently amended) A The LNAA supplement according to claim 37, wherein the LNAA supplement is substantially free from phenylalanine.

41. (Currently amended) A method for treating a subject suffering from ~~a condition involving~~ a metabolic disorder involving the metabolism of a first amino acid X, said method comprising the steps of:

obtaining a composition which is substantially free from said first amino acid X, and said composition comprises a second

amino acid Y that competes with amino acid X at a gastrointestinal tract transporter; and

enterally administering to the subject a said composition ~~which is substantially free from said first amino acid X, and which said composition comprises a second amino acid Y that competes with amino acid X at a gastrointestinal tract transporter.~~

42. (Currently amended) A The method according to claim 41, wherein the condition metabolic disorder is not phenylketonuria and/or phenylalanemia.

43. (Currently amended) A The method according to claim 42, wherein the gastrointestinal tract transporter is a Caco-2 cell transporter.

44. (Currently amended) A The method according to claim 41, wherein the gastrointestinal tract transporter is a Caco-2 cell transporter.

45. (Currently amended) A The method according to claim 44, wherein the condition metabolic disorder is tyrosinemia; ~~wherein~~ and the first amino acid X is tyrosine, and ~~wherein~~ the second amino acid Y is selected from the group consisting of Phe, Leu, Trp, Lys, His, and combinations thereof.

46. (Currently amended) A The method according to claim 44; wherein the condition metabolic disorder is tyrosinemia; ~~wherein~~ the first amino acid X is selected from the group consisting of phenylalanine, tyrosine, and combinations thereof; and wherein the second amino acid Y is selected from the group consisting of Leu,

Trp, Lys, His, and combinations thereof.

47. (Currently amended) A The method according to claim 44, wherein the ~~condition~~ metabolic disorder is alkaptonuria; wherein the first amino acid X is selected from the group consisting of phenylalanine, tyrosine, and combinations thereof; and wherein the second amino acid Y is selected from the group consisting of Leu, Trp, Lys, His, and combinations thereof.

48. (Currently amended) A The method according to claim 44, wherein the ~~condition~~ metabolic disorder is homocystinuria; wherein the first amino acid X is methionine; and ~~wherein~~ the second amino acid Y is an amino acid that competes with methionine at a gastrointestinal tract transporter.

49. (Currently amended) A The method according to claim 44, wherein the ~~condition~~ metabolic disorder ~~is a disorder~~ affecting metabolism of a branched amino acid selected from the group consisting of leucine, isoleucine, valine, and combinations thereof; ~~wherein the~~ said first amino acid X is selected from the group consisting of leucine, isoleucine, valine, and combinations thereof; and ~~wherein the~~ said second amino acid Y is an amino acid that competes with the first amino acid X at a gastrointestinal tract transporter.

50. (Currently amended) A The method according to claim 49, wherein the ~~condition~~ metabolic disorder is selected from the group consisting of maple syrup urine disease, isovaleric acidemia, methylmalonic acidemia, and propionic acidemia.

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51. (Currently amended) A The method according to claim 41, wherein said method further comprises the step of+ restricting the subject's dietary intake of the first amino acid X.

52. (Currently amended) A The method according to claim 41, wherein said method further comprises the step of—not restricting the subject's dietary intake of the first amino acid X.

53. (Currently amended) A The method according to claim 52, wherein said enteral administration is carried out substantially at mealtime.

54. (Currently amended) A The method according to claim 41, wherein said enteral administration is carried out substantially at mealtime.

55. (Currently amended) A The method according to claim 41, wherein said enteral administration is carried out orally.